Value of 99mTc-HMDP in the exploration of melorheostosis

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Citation

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Abstract

The melorheostosis is a rare osteopathy, often monomelic of chronic evolution corresponding to a osteosclerosis with hyperostosis into cast whose etiopathogeny remains unknown.
We report the observation of a melorheostosis of the lower limb occurring in a
38 years old patient. Radiography made it possible to direct the diagnosis towards the aspect of an endostosis bone condensation, into cast of candle, interesting in a unilateral way the skeleton of the left lower limb (femur and tibia). The bone scintigraphy corroborated the diagnosis by highlighting a rather intense uptake of the tracer at the level of the left femoral diaphysis exceeding the periostosis and invading the soft parts partially. A moderate uptake was also noted compared to the higher half of the left fibula. A synovectomy of the knee was carried out with double recovery. The evolution was marked by a clear attenuation of the pain.

INTRODUCTION

The melorheostosis is a rare osteopathy which etiopathogeny is still obscure, described for the first time in 1922 by Leri and Joanny like a osteosclerosis with a hyperostosis in casting. The melorheostosis prevails in a sporadic state and does not seem to take an hereditary form. We report, through this work, a rare case of this pathology.

CASE REPORT

A 38 years old patient without particular antecedent having consulted for a painful tumefaction of the left lower limb answering partially to analgesic treatment with stiffened knee in light inflection.

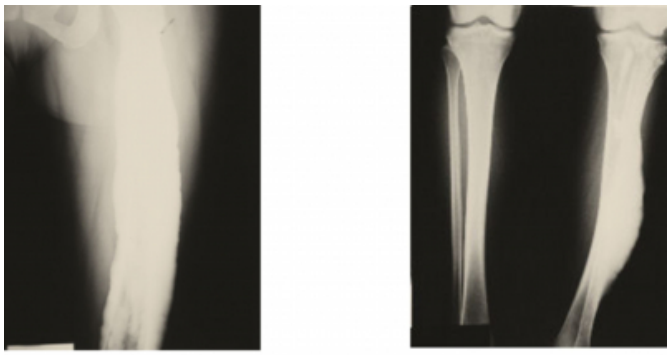
HAUT DU FORMULAIRE

The clinical examination objectified a clear asymmetry of the two lower limbs at the expense on the left side with a tumefaction prevailing in the level of the left thigh and leg. The examination of the left knee reveals a very painful inflection, limited to 70°. The remainder of somatic examination was without particularity characteristic.

Radiography showed an osseous condensation interesting endostosis with thickening of external cortical, in run candle, on all the femoral diaphysis and the higher half of the left fibular diaphysis (Figures 1).

Figure 1

Figure 1: Radiography showing an osseous condensation interesting endostosis with thickening of external cortical, in run candle, on all the femoral diaphysis (A) and the higher half of the left fibular diaphysis (B).

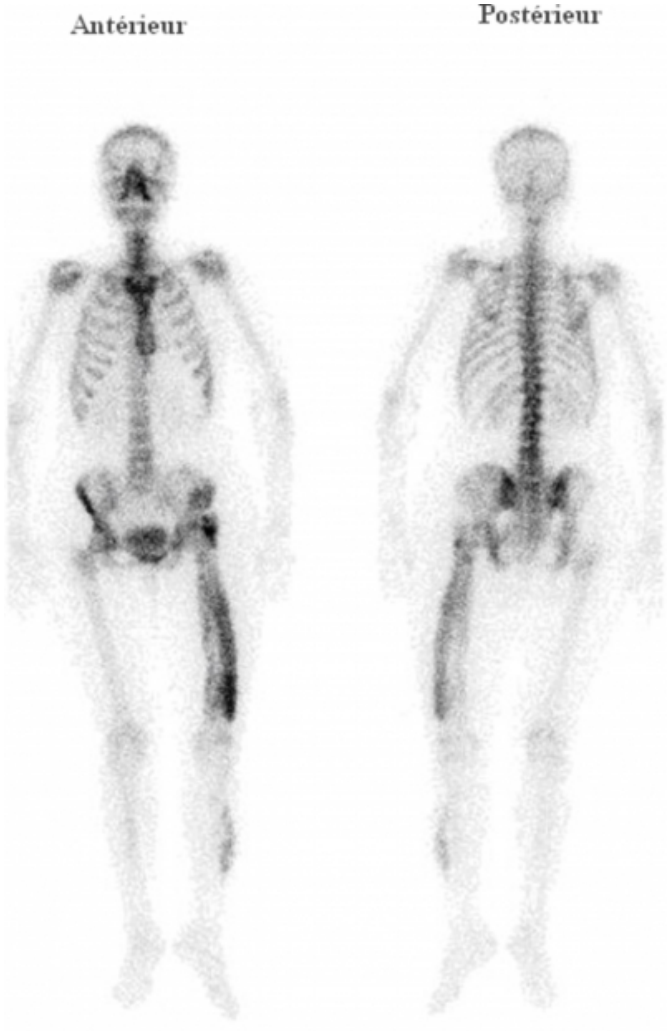


(A) (B)

The bone scintigraphy corroborated the diagnosis by highlighting a rather intense uptake of the tracer at the level of the left femoral diaphysis exceeding the periostosis and invading partially the soft parts. A moderate uptake was also noted in front compared to the higher half of the left fibula (Figure 2).

Figure 2

Figure 2: Whole body bone scintigraphy using Tc-HMDP showing an intense uptake of the tracer on the level of the left femoral diaphysis exceeding the periostosis and invading partially the soft parts with a moderate uptake in the higher half of the left fibula.



A synovectomy of the knee with biopsy were carried out with double recovery. Histologic examination showed a hyperostosis and an osteosclerosis without any specificity. The evolution was marked by a clear attenuation of pain with the persistence of a limitation of inflection of the left knee.

DISCUSSION

The melorhestosis is a rare nonhereditary and congenital anomaly often monomelic with a chronic evolution, described for the first time in 1922 by Leri and Joanny [1]. Its etiology remains unknown. The classically allowed theory supposes an early embryonic infection of a sensory nerve inducing the changes in the corresponding sclerotom, but a mosaicism could be a better explanation for sporadic forms and the asymmetrical distribution of the anomalies [2]. This pathology can appear at any age at the two sexes, with a young adult predilection. Its beginning is insidious with a progressive deformation of the ends, pains, stiffness, tumefaction of the members and limitation of movement of the articulations. These symptoms usually appear at the end of childhood or the beginning of adolescence and progress to adult age. The localizations with the lower limbs are more frequent than those of the upper limbs [3].

Radiological aspect is characterized by a cortical hyperostosis, generally only in one bone side, in “run of wax” as on a candle. Theoretically, there is quite distinct demarcation between affected and normal bone. One can also see linear densities of sclerosis especially on the level of the cortex but being able to extend to spongy bone. While especially affecting the long bones of the upper limbs and lower, the melorheostosis reaches also the short bones of the hand and the foot, but seldom the rachis. It can coexist with osteopoikilosis, striated osteopathy, tumours and vascular or lymphatic malformations. One often sees ossifications of soft tissues on the level of the articulations. Bone scintigraphy contributes as a functional exploration means, to the early detection of the osseous lesions with a more precise delimitation of their extent. They are generally images of moderate uptake of tracer interesting the cortical one preferentially taking a considerable share of diaphysal structures. Small hearth of uptake can be sometimes detected on the level of the articulations or the soft tissues as it is the case in our patient [4].

Computed tomography and resonance magnetic imagery can also contribute to the diagnosis one highlighting typical lesions of this disease. In its rough form, the melorheostosis can simulate other diseases like the ossifying myositis and osteoma. On the other hand no case of degeneration was described and even an observation of a mysterious partial regression was reported by Kanis [5].

The forecast of the disease is strictly functional. It is related to the extent of lesions, the deformations, the child growth disorders, the stiffness and articular ankylosis. More rarely at adult nervous compressions by osseous hypertrophy can be observed [6, 7].

The mysterious pathogenesis of this affection makes from there the treatment primarily symptomatic. Some treatments by radiotherapy or by diphosphonates

[8] or sympathectomy [9,10] were tested. The recourse to the

surgery is considered only in some cases where the melorheostosis becomes debilitating particulary in the event of deformation, stiffness or articular ankylosis.

CONCLUSION

The melorhestosis is a rare nonhereditary and congenital anomaly often monomelic with a chronic evolution. . Its beginning is insidious with a progressive deformation of the ends being able to evolve to a limitation of articulations movement. Bone scintigraphy contributes as a functional exploration means to the early detection and treatment of the osseous lesions.

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